APPENDIX VERMIFORMIS DUPLEX

By A. J. E. CAVE

Despite the notorious range of variation in characters and position natural to the human vermiform appendix, the extremes of variation—absence and duplicity—affect this organ so very rarely that every authentic example of either is worthy of record, particularly since such variations have been doubted or denied by past writers. The evidence set forth below, and submitted as additional material for the basis of a study of the underlying problem of causation, establishes beyond dubiety the morphological entity of these conditions, besides adducing a plausible embryological explanation of the cases of complete duplicity. The comparative anatomy of the caeco appendix is briefly reviewed to provide a suitable background for the consideration of the conditions occasionally obtaining in the human subject.

I. COMPARATIVE ANATOMY

Though the distinctive type of caecum with a true vermiform appendix is characteristic of the gibbons, Anthropoids and Man only, yet many sub-Primate forms exhibit a contracted caput caeci, associated more or less with the submucosal presence of lymphoid follicles of varying size, and descriptively at least termed an appendix. It must at once be emphasised that the criteria of such an organ differ somewhat in different authors' hands; that in atonic or long-preserved autopsy material it is not always easy to be sure of the existence *in vivo* of any functional appendix; that individual variation seems to play a definite role in some forms, and that it is often a moot point whether the modified caecal extremity of the lower Mammal is at all strictly comparable with the human or anthropoid vermiform process.

The presence, form and features of the caecum are largely determined by the nature of the particular diet, minor structural modifications being dependent upon peculiarities of intestinal arrangement and modes of mesenteric fixation.

In many Amphibia there occurs a pouching of the large gut at its reception of the small bowel, together with a local aggregation of lymphoid tissue. In certain Lacertilians paired caeca occur, or (as in *Iguana*) the caecum manifests a symmetrical pouching. Wood Jones (1912) has tentatively suggested that the primitive pro-mammalian caecum was a paired structure, and that the subsequent greater specialisation of the mammalian large intestine led to an asymmetry of its disposition, a shifting and kinking of the ileo-caecal angle, with the ultimate obliteration of the one and the persistence of the other of the original caecal pair. With Chalmers Mitchell (1905) he would regard the common

mammalian type of caecum as a unilateral development of the early reptilian and pre-mammalian condition of parts, seeking substantiation for this view in the presence of the double, bilocular, or subbifid caecum appearing in various Edentates, Sirenia, Rodents, and Ungulates. Similarly, on this hypothesis, the rudiment of one or other of the original pair of caecal pouches is said to be recognisable in Monotremes, Marsupials, Edentates and some Ungulates. This attractive speculation, receiving a certain support from zoology and embryology (*vide infra*), cannot be followed further here. Suffice it to say that the greatest caution is essential if one is not to endow the ordinary mammalian caecal sacculation with a morphological value in excess of its proper intrinsic nature, and that careful examination of a greater range of fresh material is desirable before the advancement of a decided opinion on this point.

The paired caecal appendages of the highly specialised Birds need small comment; assuming the form of small pouches in the raptorial species they appear as long tubular structures in the graminivorous kinds, thus affording a striking parallel to the brevity of the mammalian caecum in carnivorous speciës and its length and complexity in the non-carnivorous types.

Among Monotremes, *Ornithorhynchus* possesses, at the ileo-colic junction, a tubular free structure generally called caecum, provided with its own mesentery and containing much lymphoid tissue, but not differentiated into caecum proper and appendix. If a well-developed caecum be considered wanting, then this organ is the equivalent of a genuine appendix. According to MacKenzie (1916) a definite caecum and appendix occur in *Echidna* (*Tachyglossus*).

Among Marsupials the caecum is a simple undifferentiated sac (*Phalangista*, *Macropus*, *Helmaturus*, *Hypsiprymnus*, *Petrogale*, *Didelphys*), but in the wombat (*Phascolomys wombat*, *P. mitchelli*) a definite vermiform appendix is present and is subject to considerable individual variation. A similar appendix caeci is reported in *Phascolarctos*.

Among Edentates, *Orycteropus* may manifest a diminished calibre of the distal caecum; in pangolins and sloths the caecum itself is wanting; paired caeca characterise *Tatusia*, *Dasypus*, *Xenurus*, *Myrmecophaga* and *Cyclothurus*. The Cetacean caecum is a simple, bluntly rounded sac. Of the Sirenians, *Manatus* displays the paired or bifid caecum, whilst *Halichore* possesses a simple sac; neither exhibit any appendical formation.

The Ungulate caecum generally lacks any terminal differentiation, though such has been described in *Tapirus americanus*, *Babirusa*, and *Dicotyles*; in the buffaloes a subbifid caecum may occur.

An appendix in the form of a long tapering and constricted terminal caecum is present in several Rodents (*Castor*, *Myopotamus*, *Cercolabes*) but wanting in many others (*Dasyprocta*, *Hystrix* and the Sciuridae). In the hare a medial diverticulum is present additionally to the apical appendix. Probably no one of these rodent appendices is strictly comparable with the anthropoid and human appendix.

In the Carnivora the short simple caecum is usually devoid of structural

differentiation and not even a pseudo-appendix occurs in most forms: (*Canus* (dog, fox, wolf, jackal, fennec), *Nyctereutes*, *Hyaena*, *Genetta*, *Viverra*, *Suricata*, *Felis* (lion, tiger, leopard)). A descriptive appendix has, however, been reported in *Herpestes*.

The Chiroptera lack a caecum, as does *Erinaceus* among Insectivora; the Tupaiidae manifest no caecal differentiation.

The Lemuroid caecum is sacculated in *Galago*, *Loris*, and *Perodicticus*, simple and globular in *Chirogale*; its terminal portion exhibits some degree of anatomical distinctness in *Galago*, *Perodicticus* and *Chiromys*; *Nycticebus* and *Lemur* may show a well-developed appendix which is totally absent in *Tarsius* and *Loris*. An appendix has been described in *Hapale peninicillata*, in *Ateles*, and in *Lagothrix*, but *Cebus*, *Pithecia* and other New World monkeys are devoid of any such organ. In many Platyrrhine forms the termination of the conical caecum may taper to a "pointed extremity which may be elongated and resemble a vermiform appendix" (Le Gros Clark, 1934).

The absence, however, of lymphoid tissue from these so-called appendices has led Johnston (1920) to conclude that these terminal caecal prolongations are not of the nature of a true vermiform appendix.

In the Old World monkeys (*Macacus*, *Cercocebus*, *Papio*, *Cercopithecus*, *Semnopithecus*) the bluntly rounded or sometimes sacculated caecum lacks an appendix, although Wood Jones (1929) has figured it as present in a specimen of *Cercopithecus tantalus*.

The gibbons, Anthropoids and Man are characterised by the possession of a long and truly vermiform appendix arising from a capacious rounded caecum, and exhibiting a uniformity of structure, function and pathology throughout this group. Whilst the question of homologies must remain open, it is perhaps safer, in the present unsettled state of our knowledge, to regard this highest Primate type of appendix as an exclusive specialisation, lacking strict parallel among lower forms with the possible exception of Monotremes and some Marsupials.

II. CONGENITAL ABSENCE OF APPENDIX IN MAN

Two cases of total congenital absence of both caecum and appendix have been recorded, both from female subjects, the one by Robinson (1895), the other by Elliot Smith (1904). Absence of the appendix might be expected as the invariable concomitant of absent caecum were it not for the extraordinary case reported by Mouchet (1929) of a 80 years old woman in whom failure of growth and differentiation of the main caecal sac had not prevented the complete development of a typical vermiform appendix, by which alone was the first part of the large gut represented.

Cases of true congenital absence of appendix are excessively rare, as Table I indicates, only one solitary example being encountered in a specific search of over 4000 abdomens. Indeed the very existence of the condition has been denied by some (e.g. Berry, 1895), whilst Kelly and Hurdon (1905), themselves

quoting no personal case, regard all reported instances as merely examples of a secondary atrophy and obliteration, and not as genuine instances of developmental arrest.

Fawcett and Blachford's specimen was not, as Berry (1907) supposed, the sole example recorded to date, previous instances being already on record. According to Gladstone (1915) the condition was known to Meckel, and Bryant (1893) and Zuckerkandl (1894) had noted its occurrence, whilst in 1903 Huntington had published descriptions of his two specimens, the first, "a caecum having a blunt conical form without appendix", the second, a "caecum having a rounded symmetrical form, with the longitudinal muscular bands converging towards its apex, but without appendix". Gladstone himself (1915) carefully figured and described a new specimen, obtained from an elderly dissecting room female, and manifesting an "asymmetrical form of caecum without appendix".

Table I

Author	No. cases examined	Appendix absent
Treves (1885)	100	0
Ribbert (1893)	161	0
Fawcett and Blachford (1900)	350	1
Monks and Blake (1902)	641	0
Berry (1907)	100	0
Gladstone and Wakeley (1924)	3000	0
Total	4352	1

More recently Green and Ross (1933) encountered the same condition in an adult male subject, whilst Feldman (1934) recorded a right-sided caecum without appendix in a girl of 12 years, the subject of partial transposition of viscera. Though this last clinical case seems well enough founded it must be borne in mind that Bryant found an appendix of only 6 mm. in length, and Huntington one of but 5 mm.; so small an organ might easily be overlooked at operation, particularly if masked by fat, and consequently all reports from the operative field require careful scrutiny before acceptance.

To the foregoing well-established cases may be added the only specimen of its kind in the Museum of the Royal College of Surgeons of England, mentioned by Gladstone (1915) and illustrated here (see fig. 1). This specimen (Teratological Series, 549, II) is from a new-born child, the victim of epicephalocele, wherein the terminal ileum ascends parallel to and in contiguity with the medial caecal wall prior to effecting its union with the large bowel. Externally there is no demarcation of colon from caecum, nor any vestige of a caecal appendage, past or present. The case appears to be clearly one of primary congenital absence of appendix consequent upon failure of the normal caecal differentiation.

That a secondary congenital absence of appendix is, as Kelly observed, a not unknown phenomenon, seems clearly indicated by another specimen (Teratological Series, 549) in the same Museum, one collected by Sir John Bland Sutton and described in the Catalogue (and hence accepted by previous

writers) as "part of the ileum and caecum of a human foetus in which the appendix is absent and the caecum very short". Herein the ileum enters the caecum at the most dependent portion of that sac, there being no caecal bulge below the level of union of the two. But from the antero-medial aspect of the caecum an atrophied stalk-like structure ascends obliquely across the ileo-caecal junction to lie with its bulbous distal extremity closely applied to the ileal mesentery. This shrivelled pedunculated structure is undoubtedly the degenerated appendix itself, and its condition recalls very vividly those similar cases of non-pathological atrophy of this organ described by MacKenzie (1916) in the wombat (*Phascolomys mitchelli*). Some care must therefore be exercised in the examination of such specimens before a definite diagnosis of primary congenital absence is made.

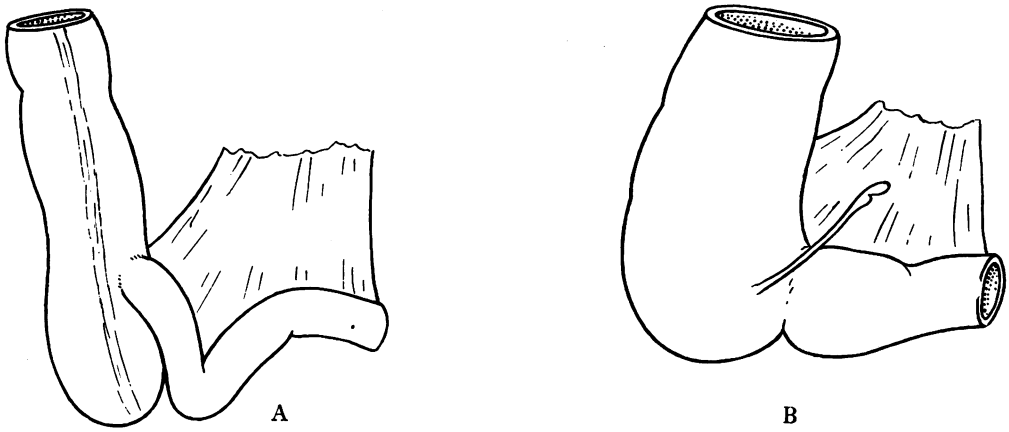

Fig. 1. A. Primary congenital absence of vermiform appendix. Mus. Roy. Coll. Surg., Teratol. Ser., No. 549, II. B. Secondary absence (congenital atrophy) of appendix. Mus. Roy. Coll. Surg., Teratol. Ser., No. 549.

One must suppose that the condition is due simply to a failure of differentiation of the primitive caecal diverticulum which makes its appearance, about the fifth week, upon the caudal limb of the extra-embryonic mid-gut.

III. DUPLICITY OF VERMIFORM APPENDIX IN MAN

Partial or complete duplicity of the vermiform appendix, though again an extremely rare condition, is nevertheless more frequently met with than the opposite extreme of total absence of that structure. The literature to date has been summarised by Greig (1934), whose excellent digest has been freely drawn upon here. Additional instances to those already published might be cited did they not emanate from the relatively uncritical field of operative surgery and in consequence lack the requisite criteria of proof.

Descriptively, known examples of appendix duplex may be resolved into the following three types:

Type A. Single caecum with one appendix exhibiting partial duplicity.

Type B. Single caecum with two obviously separate appendices (complete duplicity).

Type C. Duplicity of caecum, each caecum bearing its proper appendix.

The simplest illustrations of the first type are those curious specimens of "double-barrelled" appendix wherein the single organ presents two distinct lumina throughout either its entire length or throughout only a part thereof. Rosenberger (1903) has described such an example from an adult male, the two lumina sharing a common muscular and serosal investment. Prentiss (1907) apparently encountered a similar specimen, though its structural details are not forthcoming. Later, Elwyn (1924) discovered at operation a two-limbed appendix which he regarded as representing two originally entire organs which had subsequently undergone fusion of their distal extremities. Proximally each limb of the anomalous formation manifested characteristic appendical structure; pathological changes were absent; distal to their point of fusion both limbs shared a common lumen. A curiously divided appendix was found by Walthard (1931) in a female infant. After a normal single origin from the caecum, the structure divided into two parallel tubes which, sharing a common longitudinal musculature, reunited distally to re-establish their original common lumen.

More recently Clavel and Colson (1933) encountered an extraordinary double appendix in a thirty years old woman. Two contiguous appendices of equal length and calibre, and each manifesting the typically normal histological structure, arose together from the caecum and proceeded distally some 70 mm. Their separate and distinct tips gave the macroscopic appearance of a bifid appendix, but apart from a common peritoneal investment each organ was a distinct entity. Whether their specimen represents the ultimate stage of development of the bilocular condition exhibited by the cases just cited, or whether it is here a question of the subsequent juxtaposition of two originally separated processes, is not easily determined; if the latter view be held concerning this anomaly then the case should be transferred to type B.

It is more than likely that the non-pathological diverticulum of the appendix described by Walmsley (1929) represents an attempted bifurcation or duplicity of this organ and belongs to the first type described here. The writer has seen a very similar case secured at operation and unfortunately not preserved, wherein the diverticulum exceeded in proportions that figured by Walmsley, and the whole appearance of the specimen was that of a bifid appendix.

Of the specimens grouped under type B the earliest undoubted example was that recorded by Paterson and Emrys-Roberts (1906), where in a full-time foetus the subject of ectopia viscerum, spina bifida, and other congenital anomalies, a "small sacculated and curved appendix" lay on each side of the ileo-caecal junction. Subsequent cases occur in the clinical rather than the anatomical literature. Thus Schooler (1907) encountered at operation a 100-mm. normal appendix accompanied by a second and smaller fellow, which arose from the caecum some 20 mm. below the first, and which had perforated as the result of acute inflammation.

Young (1911) likewise met with two almost equally long appendices arising some 30 mm. apart from the caecum of a 21 years old female, each possessing its own mesentery and each the seat of purulent inflammatory change. According to Bérard and Vignard (1914) Jalaquier had operative experience of a double appendix, whilst Braatz (1929) appears to have encountered another example in a young adult female. In a 14 years old boy Goldschmidt (1930) found a supernumerary appendix arising from the anterior wall of the caecum.[1]

To these recorded instances may be added the present example which, like the first case quoted, hails from the anatomical laboratory and with that case establishes beyond cavil the entity of appendix duplex. The specimen (fig. 2)

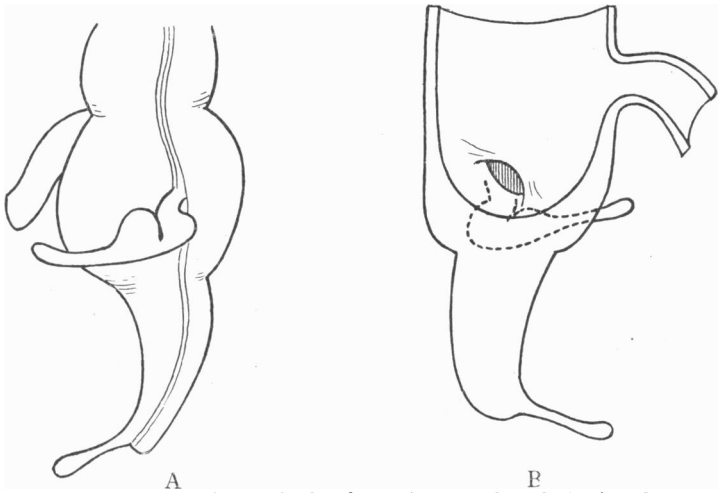

Fig. 2. Appendix vermiformis duplex from (A) ventral, and (B) dorsal aspects.

was obtained some years ago from an otherwise unremarkable full-time foetus by Mr H. C. Wilson, Prosector to the Royal College of Surgeons' Museum. The terminal ileum enters the large bowel at the customary angle, and the caeco appendix retains its infantile form. The caecum proper comprises a proximal dilated moiety succeeded immediately by a constricted conical distal moiety, from the left side of the apex of which a 15-mm. long vermiform appendix proceeds infero-medially. This appendix has now lost any original mesentery it may have possessed; it communicates freely with the caecal cavity and does not appear to be furnished with a valve of Gerlach. On the postero-medial wall of the proximal caecal segment an orifice, guarded by prominent crescentic folds of the mucosa, leads into the lumen of a second vermiform appendix. This supernumerary appendix, bound to the caecum by a short mesentery and angulated soon after its commencement, is some 20 mm. long; it has a calibre at least twice that of the apical appendix, and, save for its tip, is entirely retro-

[1] Dr D. E. Robertson, of Toronto, informs the writer (12 November, 1935) that he has seen in Boston "a specimen of a caecum that has two vermiform appendices". This case is described by Dr H. M. Pratt in the *Amer. J. Dis. Child.*, June 1933, vol. XLV, pp. 1263–76.

caecal. One of the taenia coli may be traced to the roots of both appendices, which lie about 15 mm. apart. Both structures manifest an identical normal histology.

In the interpretation of this interesting specimen the apical vermiform process is regarded as the equivalent of the normal appendix, which at this age and for some time thereafter displays such a terminal disposition. The retrocaecal appendix is supernumerary, and its presence is to be explained by the persistence and development of a fugitive embryological structure, of great morphological interest, but one commonly ignored in standard embryological works. This structure is the "transient appendix" which Kelly and Hurdon (1905) described in their study of the developing caeco appendix in over fifty young human embryos from the Mall and Brödel collections. They figured (*op. cit.* p. 72, fig. 28) this curious and temporary outgrowth from the tip of the

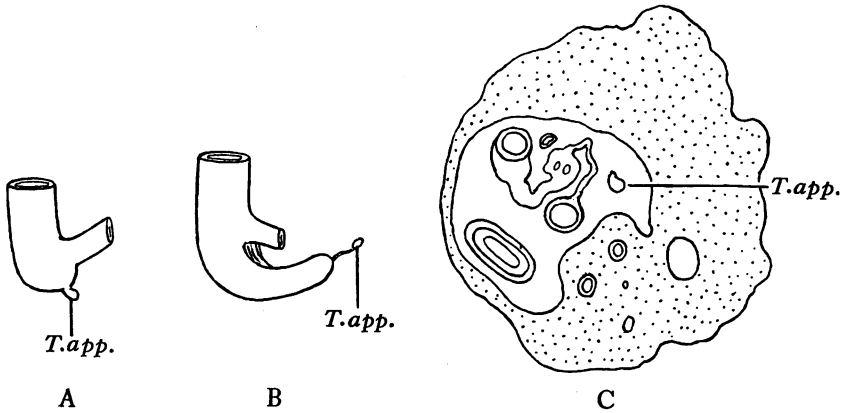

Fig. 3. The "transient appendix", (A) at the sixth week, (B) at the seventh week (after Kelly and Hurdon), (C) at the 20-mm. stage (after Gladstone).

caecum, growing during the sixth week and atrophying towards the end of the seventh week or very soon after (see fig. 3). Gladstone (1924) confirmed the presence and nature of this "transient appendix" in 10–20 mm. embryos, and has figured its appearance in a cross-section of the umbilical coelom at the 20-mm. stage. He describes this curious process as "a remarkable outgrowth from the tip of the caecum... which simulates in its position and form the true vermiform appendix", but adds, rightly, that, "since it atrophies in embryos of 20 mm. length, and afterwards completely disappears, it is believed to be an independent structure, and not connected with the permanent appendix caeci, which is differentiated later". This clear statement requires no elaboration. Suffice it here to remark that in the "transient appendix" lies a definite potential embryological origin for the development of a supernumerary appendix, and the most plausible explanation of the specimen of appendix duplex herein described as well as of the cases quoted from the literature. Whether the presence of the "transient appendix" may be regarded as affording substantiation of the theory of an ancestral caecal duplicity in the Mammalia is a

morphological question beyond the scope of this paper; for its solution there is not, as yet, a sufficiency of data from comparative vertebrate embryology.

Under type C two cases may be quoted for the sake of completeness. In a 10 weeks old female Greig (1934) found the whole bowel duplicate distal to the site of Meckel's diverticulum; two separate caeca were present, each bearing its proper vermiform appendix.

Fig. 4. Duplicity of ileum, caecum and appendix.
Mus. Roy. Coll. Surg., Teratol. Ser., No. 548, D *a*.

A specimen (fig. 4) from the Teratological Series of the Royal College of Surgeons' Museum (No. 548, D*a*) illustrates a not dissimilar duplicity of the parts derived from the apex of the primitive mid-gut loop, wherein the caecum and appendix are necessarily involved. The specimen is from a new-born infant, the subject of umbilical hernia. The ileum bifurcates so that its last 17 cm. or so consists of two separate and symmetrical tubes, each of which joins a separate caecum. The caeca are adherent mesially and each is provided with its own vermiform appendix; distally they unite to form a single ascending colon completely divided by an internal septum of mucous and submucous tissue.

Concerning such bizarre duplicity of the bowel no very satisfactory embryo logical explanation is as yet forthcoming, and further examples need not therefore be quoted.

SUMMARY

Though a remarkably constant structure in Man, the vermiform appendix is nevertheless occasionally subject to the extremes of variation, i.e. total suppression and duplicity. Cases of appendix duplex are described and are seen to fall into two categories: (*a*) supernumerary appendix due to persistence of a transient embryological structure; (*b*) appendical duplicity incidental to a more general affection of the primitive mid-gut.

REFERENCES

BÉRARD, L. and VIGNARD, P. (1914). *L'Appendicite*, p. 6. Paris.
BERRY, R. J. A. (1895). *Anat. Anz.* Jahrg. Bd. x.
—— (1907). *Intercolon. med. J. Aust.* vol. XII, p. 305.
BRAATZ, E. (1929). *Zbl. Chir.* Bd. LVI, S. 1346.
BRYANT (1893). *Ann. Surg.* vol. XVII, p. 164.
CHALMERS MITCHELL, P. (1905). *Trans. zool. Soc. Lond.* vol. XVII, pt. 5.
CLAVEL, C. and COLSON, P. (1933). *Lyon chir.* t. XXX, p. 174.
ELLIOT SMITH, G. (1904). *J. Anat. and Phys.* vol. XXXVIII, p. 32.
ELWYN, A. (1924). *Anat. Rec.* vol. XXVII, p. 180.
FAWCETT, E. and BLACHFORD, J. V. (1900). *J. Anat. and Phys.* vol. XXXIV (N.S. vol. XIV), *Proc. Anat. Soc.* 1899, p. XX.
FELDMAN, W. M. (1934). *Proc. R. Soc. Med.* vol. XXVIII, p. 753.
GLADSTONE, R. J. (1915). *J. Anat. and Phys.* vol. XLIX (3rd Ser. vol. X), p. 414.
GLADSTONE, R. J. and WAKELEY, C. P. G. (1924). *Brit. J. Surg.* vol. XI, p. 503.
GOLDSCHMIDT, W. (1930). *Zbl. Chir.* Bd. LVII, p. 3123.
GREEN, S. J. and ROSS, W. J. (1933). *Ann. Surg.* vol. XCVIII, p. 316.
GREIG, D. M. (1934). *Edinb. med. J.* N.S. (4th), vol. XLI, p. 277.
HUNTINGTON (1903). *Anatomy of Human Peritoneum and Abdominal Cavity.*
JOHNSTON, T. B. (1920). *J. Anat.*, Lond., vol. LIV, p. 66.
KELLY, H. A. and HURDON, E. (1905). *The Vermiform Appendix and its Diseases.* Philadelphia and London.
LE GROS CLARK, W. E. (1934). *Early Forerunners of Man*, p. 204. London.
MACKENZIE, W. COLIN (1916). *Lancet*, Jan. 22.
MONKS and BLAKE (1902). *Boston med. surg. J.* (Nov.).
MOUCHET, A. *et al.* (1929). *Ann. Anat. path. méd.-chir.* t. VI, p. 352.
PATERSON, A. M. and EMRYS-ROBERTS, E. (1906). *J. Anat. and Phys.* vol. XL, p. 338.
PRENTISS, E. C. (1907). *Wash. med. Ann.* vol. V, p. 25.
RIBBERT (1893). *Virchows Arch.* Bd. CXXXII, S. 66.
ROBINSON (1895). *Med. Rec.*, N.Y., vol. XLVIII, p. 757.
ROSENBERGER, R. C. (1903). *Proc. path. Soc. Philad.* vol. XXIV, p. 206.
SCHOOLER, L. (1907). *Iowa med. J.*
TREVES, F. (1885). *Brit. med. J.* vol. I, p. 527.
WALMSLEY, T. (1929). *J. Anat.*, Lond., vol. LXIV, p. 47.
WALTHARD, B. (1931). *Dtsch. Z. Chir.* Bd. CCXXX, S. 413.
WOOD JONES, F. (1912). *J. Anat. and Phys.* vol. XLVI (3rd Ser. vol. VII), p. 207.
—— (1929). *Man's Place among the Mammals*, p. 233. London.
YOUNG, W. G. (1911). *J. Amer. med. Ass.* vol. LVI, p. 195.
ZUCKERKANDL, E. (1894). *Anat. Hefte*, Bd. IV, S. 99.